was suspected and markers revealed serum ferritin 30.000 ng/ml, fibri-
gen 250 mg/dl,Tri glycerides 350 mg/dl and soluble CD25 12.000/U/ml. Bone marrow aspiration confirmed the diagnosis.Cytomegalovirus (CMV) IgM and PCR were significantly high. With regard to all the previous clini-
cal and laboratory features,a CMV-induced secondary MAS in JSLE was 
confirmed.Patient was managed according to HLH-2004 treatment protocol 
(pulsed methylprednisolone 30mg/kg/day,cyclosporine A 6mg/kg/day, IV 
etopside 150mg/m²/and IVIG.Ganciclovir was also added (5mg/kg IV q/2h x 7 days). Patient showed marked clinical improvement together with 
significant diminution of the MAS laboratory markers to normal. Two weeks 
post discharge oral dexamethasone and cyclosporine A, the patient showed a severe clinical and laboratory MAS relapse ( serum ferritin 50.000 ng/ml). The previous treatment protocol was restarted together with adding Rituximab 375mg/m² x 3 doses two weeks apart. Patient showed marked clinical and laboratory improvement and was maintained on full dose oral prednisone and oral Cyclosporine A. Tight gradual withdrawal of oral steroids was done with close clinical and laboratory follow up.

Conclusion: JSLE is a major diagnostic conundrum in pediatrics owing to its extremely variable clinical manifestations which can mimic many com-
mon pediatric conditions(e.g.malignant disease,other auto-inflammatory con-
ditions,malignancy and any specific organ associated disease). Compared to adults JSLE has a more severe disease presentation. MAS is a life 
threatening condition that requires high index of suspicion and prompt management.In our case, controlling JSLE activity using Rituximab was significantly beneficial in arresting HLH evolution.

REFERENCES

Disclosure of Interests: None declared

AB0922 UPTO-DATE ON THE EVIDENCE BASED 
INTERDISCIPLINARY GUIDELINES FOR HENOCHE-
SCHÖNLEIN PURPURA
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Background: Henoch-Schönlein purpura (HSP) is the most common child-
hood vasculitis, it is characterized by inflammation of small vessels lead-
ing to non-thrombocytopenic purpura, arthritis/arthralgia, GI hemorrhage 
and glomerulonephritis 1. There is a very poor evidence base to guide the 
management of HSP, particularly for those with the severe forms of HSP 
nephritis, Up till now there are no EULAR or ACR recommendations for 
management of HSP.

Objectives: To set an updated interdisciplinary recommendations for the 
management of HSP.

Methods: Interdisciplinary guidelines were developed with representative 
from PRINOTO Egypt for pediatric rheumatology as well as patients’ group. 
A systemic literature analysis in MEDLINE was performed, evidence 
and recommendations were graded, an algorithm for treatment and final state-
ments were discussed in a consensus meeting (Nominal Group Techni-
que). A preliminary draft was fine tuned and discussed thereafter by all 
participants (Delphi Procedure).

Results: Consensus was reached on recommendations, including a stand-
adardized treatment strategy according to the HSP severity state in the 
inividual patient. In this updated interdisciplinary guideline for HSP; treat-
ment was tailored to the patient’s pathology and its severity (figure 1).

Criterias for admission were identified as: severe arthritis or arthralgia limit-
ing ability to weight bearing and mobilization, severe abdominal pain, GI 
hemorrhage, evidence of nephritis/nephritic syndrome or renal impairment, 
evidence of neurological symptoms. Criteria for early referral to pediatric 
nephrologist were also identified including: hypertension, abnormal renal 
function, macroscopic hematrua for 5-days, nephrotic syndrome, acute 
nefritic syndrome, as well as persistent proteinuria.

Conclusion: A total of seven evidence based interdisciplinary recommen-
dations for management of HSP have been formulated, that provide an up-to-date guidance of HSP management.

Disclosure of Interests: None declared

AB0923 EFFECTS OF EXON 10 MUTATIONS VS NON-EXON 10 
MUTATIONS ON FMF PHENOTYPE AND RESPONSE TO 
TREATMENT
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Serkan Turkucar1, Balahan Makay2, Erbil Unsal1.

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Background: Familiar Mediterranean fever (FMF) is the most common monogenic periodic fever syndrome. The MEFV gene mutation encoding the pantetheinase protein results in an uncontrolled increase in interleukin-1. Today, more than 333 MEFV mutations have been identified; however, exon 10 mutations are still seem to be best correlated with clinical 
findings.

Objectives: In this study, we aim to investigate the role of exon 10 mutations vs non-exon 10 mutations on clinical features and response to treatment in patients with FMF.

Methods: Data charts of children (n=935) with FMF from Dokuz Eylul University childrens hospital and Dr.B.Uz childrens’ hospital were reviewed. Patients were divided into two groups with regard to having

Figure 1: recommendations for management of HSP

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Disclosure of Interests: None declared
exon 10 mutation or non-exon 10 mutations. Genotype-phenotype features and response to treatment were compared.

Results: There were exon 10 mutations in 631 (67.5%) patients and non-exon 10 mutations in 304 (32.5%) patients. The follow-up period was 50 (26-83.2) months. The age of symptoms onset was significantly lower in group with exon 10 positive than compared group with non-exon 10 mutation. There was no difference between the age of diagnosis and colchicine onset and the diagnosis delay time. The symptoms of fever, chest pain, and arthritis were significantly higher in the exon 10 mutation group than compared other group. Biological agent need was statistically higher in exon 10 mutation group (4.8%) than group with non-exon 10 mutation (1.3%) (Table 1).

Conclusion: In our study, it was observed that cases with exon 10 mutation have early symptoms of disease. Fever, chest pain and joint findings were more prominent in cases with exon 10 mutation than cases with non-exon 10 mutation. Additionally, colchicine resistance should be kept in mind in cases with exon 10 mutation.

REFERENCES


Disclosure of Interests: Hatice Adiguzel Dundar: None declared, ozge aytug guncemmez Speakers bureau: Novartis, Abbvie, Ceyhun Acatir: None declared, Serkan Turkucar: None declared, Balahan Makay Speakers bureau: Enzyvant, Novartis, Roche, Abbvie, Erbil Ulus Grant/research support from: Novartis, AbbVie, Roche, Koçak Pharma, Speakers bureau: Novartis, AbbVie, Roche, Koçak Pharma.


EVALUATION OF PERIPHERAL NERVOUS SYSTEM INVOLVEMENT IN PATIENTS WITH JUVENILE SYSTEMIC SCLEOROSIS AND JUVENILE SYSTEMIC LUPUS ERYTHEMATOSUS

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Background: Juvenile systemic sclerosis (JSS) and juvenile systemic lupus erythematosus (JSLE) are rare connective tissue disorders characterized by multisystemic involvement, including gastrointestinal, cardiovascular, respiratory and nervous system complications. According to data from literature, peripheral nervous system (PNS) involvement is seen in 86.7% of adult patients with systemic sclerosis. Frequency of PNS disorders is reported as 10-86.7% in adult patients. Data on PNS involvement in patients with JSS and JSLE are scarce.

Objectives: We aimed to evaluate PNS involvement in patients with JSS and JSLE. Consequently, we sought to detect patients with PNS disorders, in order to enable early diagnosis and timely intervention.

Methods: Patients with JSS and JSLE were included in a cross-sectional study. Demographic and clinical data of patients were recorded during clinical visits. All of patients were evaluated and examined for sings of PNS involvement. In order to examine mononeuropathy, polyneuropathy and trigemino-facial involvement, all patients underwent routine nerve conduction studies (NCS), blink reflex (BR) and sympathetic skin responses (SSR) evaluation.

Results: Twenty JSS (15 (75%) female) and 18 (15 (83%) female) JSLE patients were initially included. All of JSS and JSLE patients had normal neurologic examination. NCS was normal in all JSS (20/20) and JSLE (18/18) patients. SSR was not recorded in 1 (5%) JSS and in 3 (16.7%) JSLE patients. BR was recorded in all JSLE (18/18) patients and in majority of JSS patients (19/20, 95%). According to SSR, mean latency of hand and foot was similar in both patients’ groups. Amplitude of foot response was lower in JSS patients, comparing to JSLE (Table 1). Among JSLE patients, amplitude of hand response was lower than amplitudes of foot response.

Disclosure of Interests: None declared


Scientific Abstracts

Abstract AB0924 Table 1. Blink reflex responses

<table>
<thead>
<tr>
<th>Group</th>
<th>JSLE mean latency (ms)</th>
<th>JSLE mean amplitude (μV)</th>
</tr>
</thead>
<tbody>
<tr>
<td>R1 duration</td>
<td>5.9</td>
<td>6.7</td>
</tr>
<tr>
<td>R2 latency</td>
<td>10.3</td>
<td>10.3</td>
</tr>
<tr>
<td>R2 duration</td>
<td>33.5</td>
<td>32.7</td>
</tr>
<tr>
<td>R2K duration</td>
<td>30.3</td>
<td>29.6</td>
</tr>
<tr>
<td>R2K latency</td>
<td>34.4</td>
<td>34.7</td>
</tr>
<tr>
<td>R3 presence</td>
<td>3/19</td>
<td>4/18</td>
</tr>
</tbody>
</table>

According to BR, R1, R2, R2K duration and latency were not different between right and left eyes in both patients’ groups. R3 was absent in 1 (10%) bilaterally and in 1 (5%) JSS patients unilaterally (right). R3 was absent in 4 (22.2%) bilaterally and in 2 (11%) JSLE patients unilaterally. Absence of R3 was more prominent in JSLE patients, comparing to JSS. (Table 2)

Conclusion: BR could be considered as a potential indicator of neuropa-thy in JSLE and JSS patients. Data on SSR need to be evaluated in studies with higher number of patients with juvenile-onset connective tissue diseases.

REFERENCES


Disclosure of Interests: None declared
